

# Traumatic multiple lower cranial nerve palsy: a case report and review of the literature

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**SUMMARY:** Taga-Senirli R, Bajin MD, Demir N, Akyol MU. Traumatic multiple lower cranial nerve palsy: a case report and review of the literature. Turk J Pediatr 2014; 56: 557-560.

Lower cranial nerve palsy, also known as Collet-Sicard syndrome, is a fairly rare pathology characterized by unilateral palsy of the IX-X-XI-XII cranial nerves. We report a multiple cranial nerve palsy developing after a head trauma that might have been considered negligible. A 16-year-old boy was admitted with swallowing and articulation problems and difficulty lifting one shoulder after a head trauma suffered during a football match. No pathology was revealed in the subsequent imaging. Cranial nerve palsies due to head trauma are very rare. Awareness of the possibility of such a condition, leading to early recognition and treatment, may result in significant functional recovery.

*Key words:* Collet-Sicard syndrome, cranial nerve injury, head trauma.

The incidence of head trauma in general is very high: about 1 million/year in developed countries. The majority of head injury incidents take place in men aged 15-24 years; more than 90% of these are minor traumas<sup>1,2</sup>. There is a low incidence (0.3%) of post-traumatic cranial nerve palsy in mild head trauma<sup>2</sup>. The primary causes of multiple cranial nerve palsies are tumors, vascular disease, serious trauma and infection, in that order<sup>3</sup>. Traumatic injury to the glossopharyngeal, vagus and accessory nerves is infrequent and usually follows a fracture through the jugular foramen<sup>2,3</sup>. Hypoglossal nerve palsy, either isolated or not, is seen after major head trauma, usually including cervical vertebra or occipital bone fractures<sup>2,4,5</sup>. This paper reports a lower cranial nerve palsy case developing after a mild head trauma in a 16-year-old boy, with no fractures apparent in CT imaging.

## Case Report

A 16-year-old boy presented to our clinic with swallowing difficulties and left shoulder drop. It was learned that he had a preceding history of trauma. He had experienced a head trauma two years previously during a football match. Afterward, he felt tension in his left shoulder. In the succeeding days, shoulder drop and loss

of strength, as well as difficulties in swallowing and in articulating some letters (“r” and “s”) appeared. He had no significant past medical or family history. He denied having experienced any other trauma. Pediatric neurology had examined our patient and could not find an underlying cause explaining the peripheral pathology.

On examination, his left shoulder was dropped. His ears, nose and neck movements were normal. Examination of the oropharynx and larynx revealed dysfunction of the left-side IXth, Xth and XIIth cranial nerves, with loss of gag reflex and paralysis of the soft palate musculature on the left side, a hypotonic tongue with deviation to the left on protrusion and left-sided vocal cord palsy with pooling of saliva in the hypopharynx. The rest of the neurological as well as audiologic examination was normal. Electromyography of the accessory nerve revealed nearly total axon loss. Magnetic resonance imaging (MRI) of the brain and MR angiography revealed no abnormalities. CT, with the window level set for bone visualization, revealed no fracture in the jugular foramen or any part of the skull.

The complaints were diagnosed as Collet-Sicard syndrome caused by the head trauma. Since so

much time had elapsed, no anti-inflammatory medication was given. However, the patient was scheduled for a series of physical therapy sessions due to his swallowing difficulties. No improvement in any of the clinical evidences was noted one year later, that is, the third year after the trauma.

### Discussion

Mild head traumas are usually referred to as those showing an initial Glasgow Coma Scale score of 13-15. Multiple CN injury incidence is about 4-7%, constituting 22-32% of patients whose cranial nerves are affected<sup>2,3,6</sup>. In the published literature, injuries to the sixth and seventh cranial nerves are those that most commonly follow head trauma<sup>2,3,6</sup>. Traumatic injury to the glossopharyngeal, vagus, accessory and hypoglossal nerves is rare and usually follows a skull base fracture<sup>2,4-7</sup>.

Unilateral palsy of the lower (IX-X-XI-XII) CNs is known as Collet-Sicard syndrome (CSS). This syndrome was initially reported by F. Collet (1870-1966), an otorhinolaryngologist, and J. A. Sicard (1972-1929), a neurologist<sup>4,8,9</sup>. Collet reported the first case of complete unilateral paralysis of all of the lower cranial nerves due to a bullet injury in the mastoid region<sup>4,10</sup>. Sicard described a number of cases showing similar clinical features<sup>4,11</sup>. Generally, a pathological process located near the jugular foramen results in Collet-Sicard syndrome<sup>12</sup>.

Involvement of the jugular foramen region leading to lower CN involvement is characterized by several eponymous syndromes in which multiple CN involvement is the rule. The dysfunction of CNs X-XI-XII is termed Jackson's syndrome. Vernet's syndrome is characterized by unilateral paralysis of CNs IX-X-XI; it generally arises due to a lesion inside the skull<sup>12-14</sup>. Collet-Sicard Syndrome refers to palsy of CNs IX-X-XI-XII and is a result of extracranial causes, though theoretically intracranial pathology could also occur<sup>10-12,14</sup>. When CSS is accompanied by ipsilateral Horner's syndrome, it is called Villaret's syndrome<sup>12-14</sup>.

Collet and Sicard provided the original descriptions of CSS based on post-traumatic cases during World War I, in which the location of the lesions, that is, the presence of metallic fragments, was demonstrated via radiographic

studies. Since then, many causative pathologies have been described. Tumors are the most common cause of CSS<sup>12,14</sup>. Tumors of the skull base, metastases of cancers such as those of the prostate, breast, kidney, lung, cervix and colon<sup>8,9, 12-14,19</sup>, or primary tumors such as glomus jugulare, schwannoma of the hypoglossal nerve<sup>14,15</sup> and fibrosarcoma of the neck<sup>16</sup> have been described. Vascular pathologies, such as internal carotid artery (ICA) dissection, ICA aneurism or internal jugular vein thrombosis have been described as causes of CSS<sup>12,14,17,18,20</sup>. Another relatively common cause is trauma, generally accompanied by skull base fracture. The current literature has no reported cases of mild trauma resulting in CSS. This case is interesting since it implicates a minor head trauma, with no signs of fracture or any other pathology to possibly explain the multiple CN paralysis.

A group of infectious diseases, such as Lyme disease, cytomegalovirus (CMV), encephalitis and HIV-related pathologies were evaluated in terms of differential diagnosis. Our patient was negative on serology and ELISA for *Borelia burgdorferi*, CMV and EBV. No signs of HIV or related central nervous system infections were detected. Systemic vasculitic diseases such as PAN, SLE and Wegener granulomatosis are known to show multiple cranial nerve palsies during their course. In our patient, neither serology nor radiological imaging was positive for any of these pathologies. No dural meningeal enhancement in MRI or any changes in the blood chart were noted; there were no signs of central nervous system or hematological malignancy. Diphtheria is another pathology that can develop polyneuropathy during its

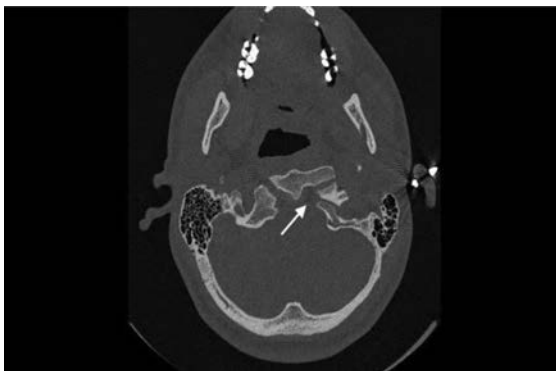


Fig. 1. Axial temporal CT image, showing normal jugular foramen

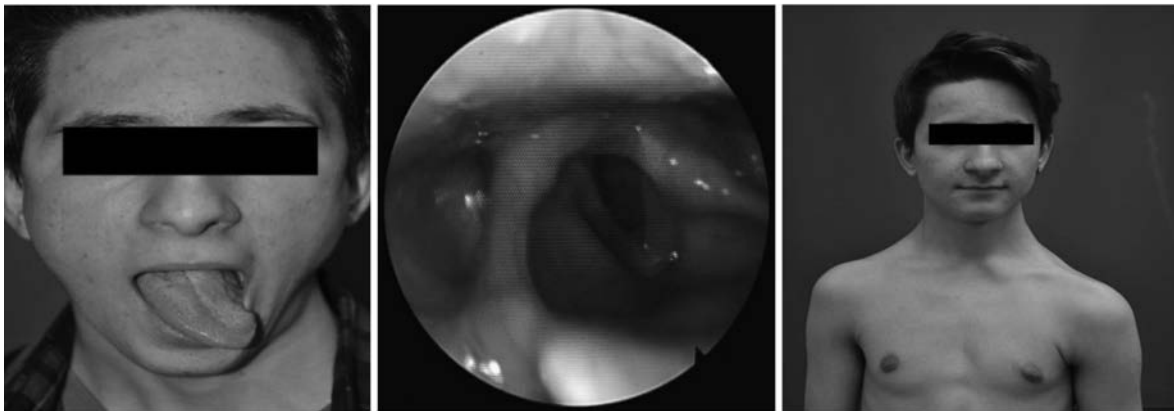


Fig. 2. Photographs of the patient showing tongue deviated to the left, left vocal cord palsy and left shoulder drop

course, but since no acute bulbar symptoms or any general demyelinating symptoms had ever been seen in our patient, this diagnosis was also ruled out.

In this case, the cause of CN IX-X-XI-XII paralysis is still to be explained. The MRI and MR angiography were negative for soft tissue or vascular pathologies, and CT imaging was negative for fractures crossing either the jugular foramen or the hypoglossal canal. We presume that the CNs were injured by soft tissue edema or a small hematoma due to minor venous bleeding within the jugular foramen and hypoglossal canal in the early post-traumatic period and a probable scarring process during late post-traumatic period. Avulsion injury to the nerves could also be the responsible pathology.

Collet-Sicard syndrome is an unusual and complex pathology. This could be an explanation for the delayed presentation and diagnosis in this case. In all cases of suspected CSS, a focal primary lesion or metastatic disease needs to be excluded initially. If malignancy is not the cause, further examination should be conducted in order to diagnose any possible fracture or vascular or infectious pathology. Although such a situation is extremely rare, if no actual lesion is found, anti-edema/inflammatory treatment should be administered and surgical exploration/decompression could be performed. This could result in functional recovery.

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